

Castleman's disease with autoimmune haemolytic anaemia, subfertility and meningomyelocele

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Introduction

Castleman's disease (CD) (angiofollicular lymph node hyperplasia) was first described in 1936 (1). It is a rare disease of unknown aetiology affecting the lymph nodes. CD consists of two basic histological types: hyaline-vascular (HV) and plasma-cell (PC), and a mixed type called hyaline-vascular plasma cell type (HV-CV) (2). Clinically, it presents as an indolent disease confined to a single site (unicentric) or less commonly as a more generalised lymphadenopathy (multicentric), accompanied by constitutional symptoms, acute phase response, organomegaly and potential for malignancy. Anaemia is common but usually mild to moderate, normochromic and normocytic (3). Bone marrow cellularity and the myeloid/erythroid ratio (M/E) are normal (3).

Haemolytic anaemia as the presenting feature of CD has been described only twice before (4,3). We report a case of CD with autoimmune haemolytic anaemia, subfertility, subsequent conception and delivery of a baby with a meningomyelocele.

Clinical presentation

A 30-year old woman was admitted with progressive breathlessness, malaise and weakness for one month. She was anaemic (Hb 3.4 g/dl) and had enlarged supraclavicular and left axillary lymph nodes. The lymph nodes were non-tender, smooth and 2 to 4 cm long. Further clinical examination was unremarkable except for a 5 cm hepatomegaly and 4 cm splenomegaly.

The ESR was 174 mm, but the C-reactive protein was <6 mg/l. Antinuclear factor was negative. The white cell count was 3800 x 10⁹ with a normal differential count. The blood picture showed polychromasia and spherocytes. The reticulocyte count was 5.6%. The direct Coomb's test was positive, confirming autoimmune haemolytic anaemia. Bone marrow biopsy showed an inversed M/E of 1:2 with hyperplastic and megaloblastic erythropoiesis, and normal granule- and thrombopoiesis. There was no evidence of non-haemopoietic cell infiltrate, and the plasma cells and lymphocytes were seen in normal numbers. The chest xray

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and serum protein electrophoresis were normal. Ultrasound scan of the abdomen confirmed hepatosplenomegaly.

Excision biopsy of the enlarged lymph nodes showed lymphoid follicular hyperplasia. The interfollicular area was densely infiltrated by plasma cells. Prominent perinodal fibrosis was seen. There was no evidence of a neoplastic lesion. The histology confirmed a diagnosis of plasma-cell type Castleman's disease.

Although the treatment of unicentric CD is excision of the offending lymph nodes, treatment of multicentric CD is unclear. Systemic therapy with steroids alone or in combination with chemotherapeutic agents and radiotherapy has met with varied success (3,6,7,8).

In consultation with an oncologist the patient was managed with prednisolone, vitamins, iron and folate supplements. Starting prednisolone at 40 mg/day for one month, the dose was tapered to a maintenance dose of 10 mg/day. She was followed up monthly at the clinic and the haemoglobin remained stable at 9 to 10 g/dl. At 6 months she became pregnant for the first time after four years of marriage. Regular assessment in a clinic with a consultant obstetrician was arranged from the start of pregnancy. The maintenance dose of prednisolone was continued. Her condition was stable till the 28th week of amenorrhoea. Thereafter the ESR started rising and she became increasingly anaemic. The fetal growth was assessed to be normal. She was admitted to the ward during the 28th and the 32nd weeks for blood transfusions. In the 34th week the baby was delivered by caesarian section because of preterm labour and meconium stained liquor, but it was found to have a meningomyelocele and paraplegia.

Discussion

CD is known to cause reversible subfertility (9). This patient has had regular sexual intercourse without contraception for four years. She was treated with corticosteroids after excision of some enlarged lymphnodes, following which the disease became quiescent for several months. She conceived during this time.

Folate deficiency is known to cause neural tube defects. The patient was on folate supplements for 6 months

before conception and the haemoglobin was stable. Prednisolone could not be discontinued considering her clinical status. In addition, there is no proven teratogenic effect of corticosteroids (10). CD has not previously been associated with fetal neural tube defects. Since it is a rare disorder, and pregnant women with CD are rarer, this may be the first case of CD associated with fetal meningomyelocele.

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